

# Monostotic Fibrous Dysplasia of the Thoracic Spine

## A Case Report

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**Study Design.** This case report details the diagnosis and treatment of a 12-year-old boy with progressive paraparesis from monostotic fibrous dysplasia of the thoracic spine.

**Objectives.** The authors discuss their experience in the context of previous reports to recommend the optimal management of this disease.

**Summary of Background Data.** Isolated vertebral involvement with fibrous dysplasia is exceedingly rare, with only 15 case reports describing the clinical presentation and treatment of these patients. The various treatments reported have included biopsy, decompression, curettage, and excision with or without fusion. A consensus for management has not been achieved.

**Methods.** The patient developed progressive neurologic deterioration from a combination of epidural extension of dysplastic tissue and a severe subluxation with kyphosis. Evaluation included plain radiographs, magnetic resonance imaging, computed tomography-assisted biopsy, and a bone scan. The patient was successfully treated with complete resection of both the bony and soft tissue components of the lesion by a single-stage combined approach involving posterior resection, instrumentation, and fusion to achieve rapid decompression and immediate stabilization followed by anterior resection with strut graft fusion to obtain a complete resection and prevent a delayed progressive kyphotic deformity.

**Results.** The patient's neurologic symptoms and signs rapidly resolved postoperatively. Graft incorporation with normal alignment was radiographically demonstrated at 3 months and maintained at 12 months. He has no pain or disability at 17 months follow-up.

**Conclusions.** The authors recommend radical removal of all involved bone accompanied by internal fixation and bone grafting to achieve long-term stabilization in patients with monostotic fibrous dysplasia of the spine accompanied by neurologic deficits, instability or progressive pain. [Key words: fibrous dysplasia, thoracic vertebrae, child] *Spine* 1996;21:860-865

### ■ Introduction

Fibrous dysplasia of bone is characterized by malfunction of bone-forming mesenchyme.<sup>14</sup> The histopathologic appearance consists of poorly organized trabeculae of immature bone and cartilage within a fibrous matrix.<sup>12,14</sup> Two clinical variants of this disorder have been described: a polyostotic form affecting multiple bones, particularly long bones, and a monostotic form affecting a single bone.<sup>2</sup> Isolated involvement of the spine is rare. Since fibrous dysplasia is a disorder of developing bone, this condition manifests most commonly during childhood or adolescence and often becomes quiescent after skeletal maturation.<sup>2,23,27</sup> The development of symptoms in adulthood generally results from weakening of bone rather than disease progression.<sup>11</sup>

Although, on histopathologic grounds, fibrous dysplasia is considered a benign disease of bone, bony involvement occasionally will progress rapidly and inexorably, leading to profound bony destruction and severe clinical symptoms. When this clinical presentation is encountered, it is imperative to recognize the disease, identify the neurologic deterioration, and administer prompt and appropriate treatment to optimize functional outcome and long-term disease control. Herein, we describe an unusual case of thoracic fibrous dysplasia and emphasize relevant factors in the management of this child that contributed to his favorable outcome.

### ■ Case Report

A 12-year-old boy of Greek descent with an unremarkable past medical history developed progressive painless lower limb weakness and numbness over several weeks. He described Lhermitte's sign with neck flexion. On admission, his gait was remarkable for significant ataxia and weakness of the lower limbs. He had marked urinary retention. Physical examination revealed no spinal tenderness to direct palpation. His spinal range of motion was not impaired. A small cafe-au-lait spot was present on the left arm. Neurologic examination showed impaired sensation below the T11 dermatome and paraparesis with ability to move against gravity but not against resistance. Bilateral lower limb hyperreflexia and extensor plantar responses were observed. Sphincter tone was normal.

Laboratory studies included a complete blood count, erythrocyte sedimentation rate, and calcium and alkaline phos-

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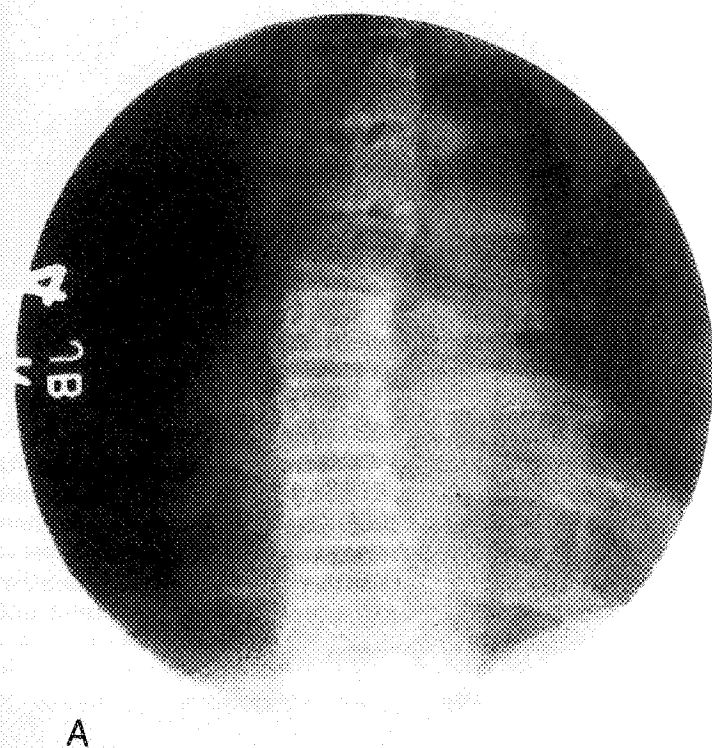


Figure 1. Anteroposterior (A) and lateral (B) plain radiographs demonstrate slight collapse and anterior translation of the fifth thoracic vertebra with erosion of the right pedicle.

phatase, which were all within normal limits. Skin testing with purified protein derivative was normal. Plain radiographs of the thoracic spine revealed collapse and 10 mm of anterior translation of the fifth thoracic vertebral body (Figure 1) signifying probable instability. Magnetic resonance imaging demonstrated diffuse vertebral body involvement of the fifth thoracic vertebral body with right lateral and posterior paravertebral and epidural extension; minimal enhancement was detected after administration of intravenous gadolinium (Figure 2). A biopsy was performed to rule out such diseases as aneurysmal bone cyst, hemangioma, or mycobacterial infection which could be treated nonoperatively. Histopathologic evaluation of a closed needle biopsy specimen obtained via computed tomography guidance revealed a moderately cellular fibrous matrix with poorly oriented bony trabeculae consistent with fibrous dysplasia (Figure 3). The axial computed tomography images confirmed the magnetic resonance imaging findings of significant bony destruction involving the vertebral body and extending to the pedicle, transverse process,

and lamina on the right side with preservation of a rim of cortical bone (Figure 4). A technetium-99m bone scan was obtained to identify other sites of involvement; isolated mid-thoracic uptake was observed, confirming the diagnosis of monostotic fibrous dysplasia of the thoracic spine.

The patient's progressive neurologic deterioration was believed to have resulted from a combination of vertebral translation and angulation and circumferential soft tissue compression of the thecal sac. The goals of treatment included decompression of the spinal canal, complete excision of the tumor, and establishment of spinal stability. It was thought that using either an anterior or a posterior approach alone would permit decompression but would not allow either complete tumor removal or adequate stabilization given the extensive three column involvement. Accordingly, it was thought that a combined posterior-anterior surgical approach would provide the best treatment by facilitating excision of the involved vertebral body, lateral and posterior elements, and abnormal soft tissue, and allowing secure anterior and poste-

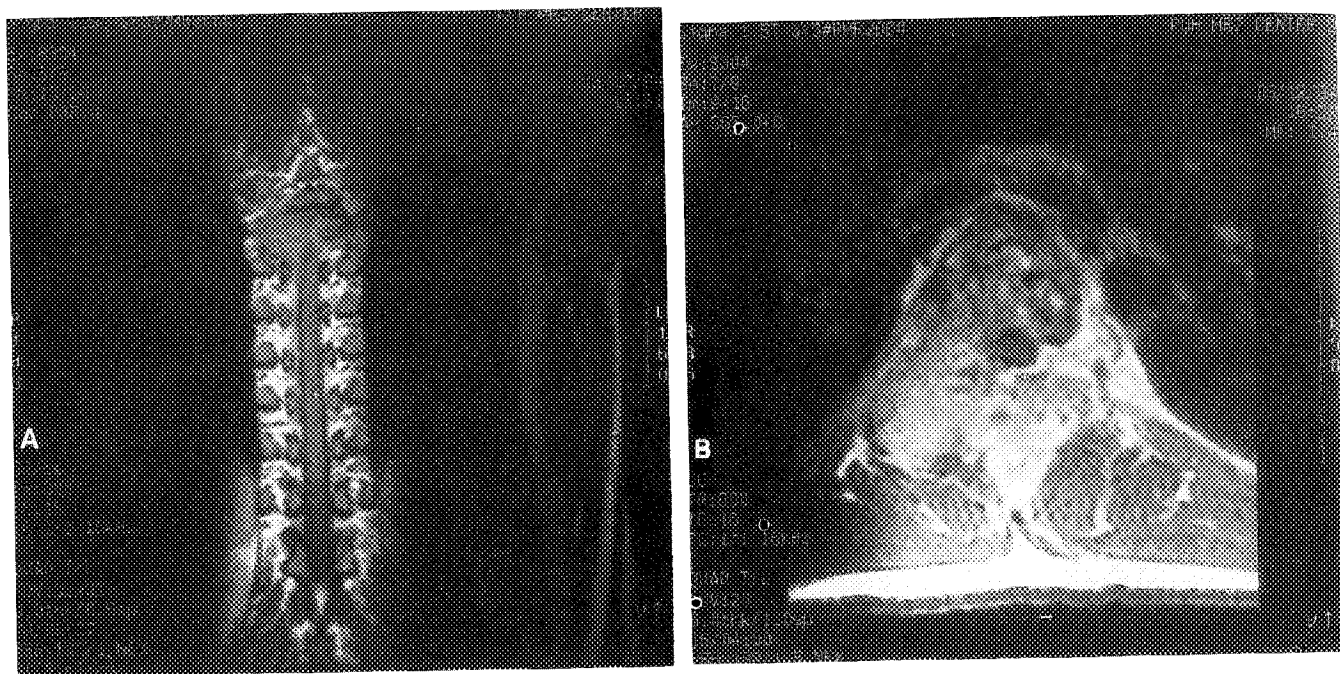


Figure 2. Coronal T-1 weighted magnetic resonance imaging (A) demonstrates epidural and paraspinous extension of the mass at the fifth thoracic segment. The gadolinium-enhanced axial T-1 weighted image (B) shows fibrous dysplasia involving the body, right pedicle, transverse process, and lamina with epidural and paraspinous extension.

rior fixation with bone grafting to prevent a delayed kyphotic deformity. The posterior approach was performed first to achieve rapid decompression and immediate stabilization.

Intraoperative somatosensory evoked potentials were monitored throughout the procedure. Partial reduction of the translation was achieved after positioning the patient prone. A complete laminectomy allowed removal of the epidural soft tissue component; a right facetectomy and removal of the involved pedicle and transverse process was then completed. The lateral paravertebral component of the mass, which was comprised of soft brownish tissue, was also removed. Short

segment rod fixation was performed utilizing a simple claw construct with bilateral transverse process and pedicle hooks at the third and seventh thoracic levels, and superior and inferior transverse cross-links (AcroMed, Cleveland OH). After decortication of the posterior elements at these levels, autologous iliac crest graft was harvested and placed around the construct to facilitate bony fusion.

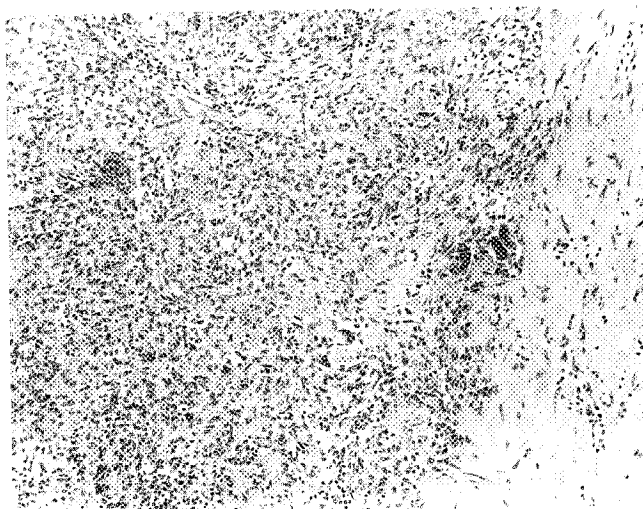


Figure 3. Photomicrograph of a hematoxylin and eosin-stained section ( $\times 100$ ) of the mass reveals typical features of fibrous dysplasia, including a moderately cellular fibrous matrix with poorly oriented trabeculae of bone.

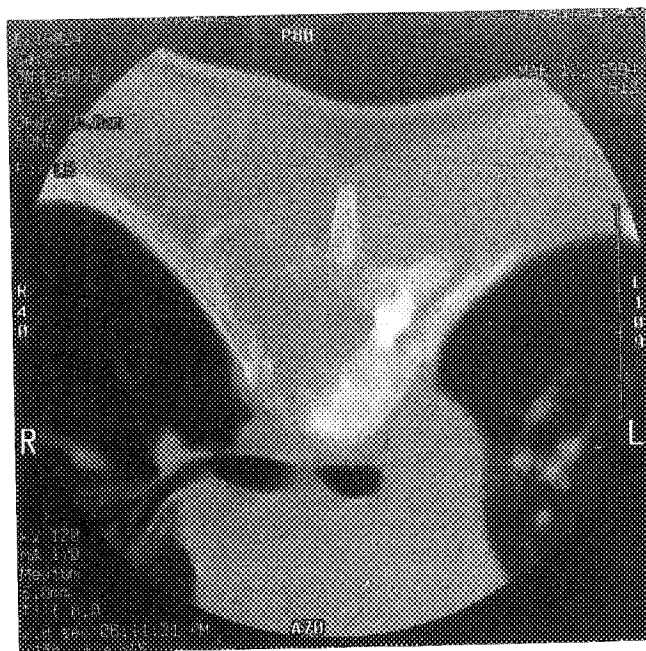


Figure 4. Unenhanced axial computed tomographic image windowed for bone demonstrates the preservation of a cortical rim of bone surrounding the destructive process of the right body, pedicle, transverse process, and lamina of the fifth thoracic segment.

**Table 1. Summary of Previous Case Reports of Monostotic Fibrous Dysplasia of the Spine**

Author	Age (yrs)	Sex	Duration*	Trauma	Region	Treatment	Outcome
Schlumberger <sup>25</sup>	20	M	11 mo	Neck	Right C4 <sup>b</sup>	Biopsy	Not given
Ledoux-Lebard and Soulquin <sup>13</sup>	58	F	6 yr	None	L1 <sup>b</sup>	Biopsy/decompression	Not given
Rosendahl-Jensen <sup>24</sup>	35	F	1 yr	Neck	Left C4 <sup>b</sup>	Curettage	No pain at 1 yr
Harris et al <sup>6</sup>	42	M	Not given	None	Right L4 <sup>b</sup>	Biopsy	Pain at 4 yr
Nyul-Toth and Joos <sup>19</sup>	26	M	3 yr	None	Right L2,3 <sup>b</sup>	Biopsy/fusion	No pain at 1 yr
Daniluk and Witwicki <sup>3</sup>	28	F	10 yr	None	Right L4 <sup>p</sup>	Excision	No pain
Resnik and Lininger <sup>21</sup>	27	F	Acute	MVA	Right C6 <sup>b</sup>	Biopsy	Not given
Rosenblum et al <sup>23</sup>	20	M	Acute	Head	Right T1 <sup>b</sup>	Excision/fusion/halo	Fused at 3 mo
Nigrisoli <sup>17</sup>	36	F	3 yr	None	L3 <sup>a</sup>	Curettage/fusion	Pain at 14 mo
Troop and Herring <sup>28</sup>	12	F	Acute	Fall	Right L3 <sup>b</sup>	Curettage/fusion/brace	No pain/fused at 3 yr
Kahn and Rosenberg <sup>12</sup>	23	M	Acute	Fall	Right L3 <sup>b</sup>	Excision/fusion/internal fixation <sup>n</sup>	Pain/fused at 9 mo
Hu et al <sup>8</sup>	41	M	6 mo	None	Right C2 <sup>b</sup>	Biopsy/fusion/internal fixation/halo	Fused at 30 mo
Singer et al <sup>26</sup>	44	F	1 wk	None	T11 <sup>a</sup>	Excision/fusion/internal fixation	No pain at 2 yr
Nishiura et al <sup>18</sup>	37	M	6 mo	None	C1-3 <sup>a</sup>	Excision/fusion/halo	Fused at 1 yr
Nabaro and Giblin <sup>16</sup>	46	F	6 wk	None	Right T7 <sup>b</sup>	Excision/fusion/internal fixation <sup>n</sup>	Fused at 6 mo
Przybylski et al (current study)	12	M	2 wk	None	Right T5 <sup>b</sup>	Excision/fusion/internal fixation	No pain/fused at 17 mo

\* "Duration" refers to duration of pain before diagnosis except for the patients described by Nishiura et al<sup>18</sup> and our patient who had initial symptoms of painless myelopathy.

MVA = motor vehicle accident. <sup>a</sup> = anterior elements only. <sup>p</sup> = posterior elements only. <sup>b</sup> = both anterior and posterior elements involved. <sup>n</sup> = new neurologic deficits and progressive pain after diagnostic biopsy.

After completion of the posterior procedure, an anterior T5 corpectomy with rib graft fusion from T4 to T6 was performed through a right thoracotomy. The T5 vertebral body was very soft and friable making decompression relatively straightforward. The estimated intraoperative blood loss was 3700 cc; the patient received a three unit blood transfusion. A thoracolumbosacral orthosis was applied postoperatively and ambulation was encouraged.

Postoperatively, the patient's neurologic status improved rapidly to normal sensation, strength, and urinary function over the ensuing several weeks. Postoperative radiographs demonstrated good spinal alignment. Graft incorporation and maintenance of alignment were seen on plain radiographs 3 months later and the orthosis was removed; graft incorporation and alignment were maintained on radiographs 1 year postoperatively. Currently, the patient is back to full activities, without pain or disability of any kind at 17 months follow-up.

## Discussion

Fibrous dysplasia is a common benign childhood disease of bone, accounting for 7% of benign bone tumors.<sup>23</sup> Other descriptive names for this disease include osteitis fibrosa disseminata and osteodystrophia fibrosa.<sup>1,9,15</sup> It is characterized by the replacement of the medullary component of one or, less commonly, several bones with fibrous tissue and irregular osteoid formation.<sup>14,23</sup> The etiology is not known.<sup>24</sup> The monostotic form is seven times more common than the polyostotic variant.<sup>26</sup> This latter form may occur as an isolated entity or in conjunction with abnormal skin pigmentation, premature sexual development, and other endocrinologic and extraskelatal abnormalities.<sup>1,9,14,15</sup> The association of polyostotic involvement with cutaneous and endocrine

manifestations is termed the McCune-Albright syndrome.<sup>1,15</sup>

Fibrous dysplasia affects males and females equally, and manifests predominantly during childhood or adolescence.<sup>23</sup> Although the disease has a familial tendency, a mode of genetic transmission has not been described.<sup>26</sup> After cessation of bone growth, fibrous dysplasia may remain quiescent or, rarely, actually regress.<sup>2,6,23,27</sup> Malignant transformation is very uncommon.<sup>10,22</sup>

Vertebral involvement with fibrous dysplasia is exceedingly rare, particularly in the monostotic variant. Several large series have failed to note a single patient with monostotic spine involvement.<sup>4,5,7,20</sup> A review of the literature reveals only 15 case reports describing the clinical presentation and treatment of patients with monostotic fibrous dysplasia of the spine.<sup>3,6,8,12,13,16-19,21,23-26,28</sup> Although Dahlin and Uni noted two additional cases of vertebral involvement in a series of 418 patients with monostotic fibrous dysplasia, clinical descriptions were not provided.<sup>2</sup> Features from the case reports are summarized in Table 1. Our own case is the first report of monostotic fibrous dysplasia involving the thoracic spine of a child. Including this case report, there were eight males and eight females, who ranged in age from 12 years to 58 years, with a median age of 26.5 years. At the time of diagnosis, 14 patients had pain, 3 had myelopathy, and 1 had radicular sensory loss. Six patients had antecedent trauma.

Fibrous dysplasia involved the cervical spine in five patients, the thoracic spine in four, and the lumbar spine in seven. Bone involvement of the affected levels tended

to be extensive: 12 of 16 patients had involvement of both the vertebral bodies and posterior elements; only four had isolated anterior ( $n = 3$ ) or posterior ( $n = 1$ ) disease. Interestingly, the right side was selectively involved in 11 of 12 patients with posterior element disease. Three patients were treated with biopsy alone, one with biopsy and decompression, two with biopsy and fusion, three with curettage debridement, and seven with excision. Ten patients underwent fusion: 5 with internal fixation, 3 with external immobilization, and 2 without either adjunct.

The rarity of this disorder and the lack of detailed reports of long-term follow-up makes comparison of the efficacy of the different treatment modalities difficult. However, there is some evidence that incomplete resection and/or inadequate stabilization may afford a less favorable outcome. Two patients had progressive bony destruction seen on plain radiographs 2 months<sup>24</sup> and 2 years<sup>12</sup> after the recognition of their bony disease but before treatment was instituted. Two patients, including the second with radiographic progression, developed increasing pain and new neurologic deficits within 6 weeks of a diagnostic biopsy, necessitating excision, fusion, and internal fixation.<sup>12,16</sup> Outcome was described in only three of six patients undergoing biopsy without subsequent tumor excision; two with fusion had no progression over 1 to 2 years<sup>8,19</sup> while the other without fusion had persistent pain after 4 years.<sup>6</sup> In contrast, outcome has generally been favorable in those patients treated with aggressive resection and fusion.<sup>12,16,18,23,26</sup>

The less favorable results after incomplete resection and inadequate stabilization likely reflects several characteristic features of this disorder. First, following incomplete resection, fibrous dysplasia may involve the adjoining bone or progress to encompass the bone graft. Second, the often extensive spinal involvement may cause spinal instability that could result in clinical deterioration.

Evaluation of outcome from treatment of extraspinal fibrous dysplasia also supports the concept of aggressive resection and stabilization in the management of this disorder. Progressive dysplastic involvement of bone grafts<sup>6</sup> or adjacent bony elements<sup>26</sup> when incomplete resection is obtained has been described. Stephenson et al<sup>27</sup> found that outcome in both conservative and operative management without stabilization was unsatisfactory in more than 80% of children. In contrast, if internal fixation was used in conjunction with open curettage and bone grafting, 86% of patients had satisfactory results.

In the context of these observations, the results in our patient provide support for aggressive resection with rigid fixation and fusion in the management of spinal fibrous dysplasia accompanied by neurologic deficit, instability, or progressive pain. By performing anterior and posterior tumor resection and fusion under the

same anesthetic, we were able to achieve rapid decompression of the spinal cord and correction of the bony deformity. This led to prompt recovery of neurologic function and return to ambulation postoperatively.

### ■ Summary

Monostotic fibrous dysplasia of the spine is rare. However, it is important to recognize this benign process to provide optimal treatment of the symptomatic patient. Although disease stabilization has been reported after incomplete resection and fusion, fibrous dysplasia can progress to involve adjacent bone by direct extension as well as result in graft destruction, leading to a poorer long-term outcome. Consequently, the authors recommend radical removal of all involved bone accompanied by internal fixation and bone grafting to achieve long-term stabilization.

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